Hyperacusis

David M Baguley MSc MBA

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Disorders of loudness perception, for long a clinical enigma, can represent a serious challenge to the patient. In this paper I review what is known of hyperacusis—the mechanisms and approaches to treatment. I begin with definitions, because even basic terminology still varies in this under-researched area.

Hyperacusis has been defined as 'unusual tolerance to ordinary environmental sounds'[1] and, more pejoratively, as 'consistently exaggerated or inappropriate responses to sounds that are neither threatening nor uncomfortably loud to a typical person'.[2] Common to both is the implication that the experience can be evoked by sounds of low intensity and that sounds in general, rather than specific sounds, are problematic. This is less true of *phonophobia* (fear of sound) and the recently proposed *misophonia* (dislike of sound),[3] both of which carry a suggestion that the intolerance may be specific to certain sounds with emotional associations. In neurology, phonophobia tends to be used specifically for the loudness intolerance reported by some patients with migraine.[4] For the wider types of hearing hypersensitivity, therefore, the term hyperacusis is preferable. *Loudness recruitment*[5,6] describes an experience commonly associated with cochlear hearing loss and specifically with dysfunction of the outer hair cells of the organ of Corti: with a rising sound level, the perceived loudness increases faster than normal.[6] This phenomenon may be distinguished from hyperacusis if the individual perceives sound of moderate intensity as uncommonly loud (recruitment) or sound of low intensity as uncomfortably loud (hyperacusis) but the two experiences are not mutually exclusive. Loudness recruitment does not, however, vary with mood.

PREVALENCE, INCIDENCE AND QUANTIFICATION OF HANDICAP

Lack of robust epidemiological data is a major shortcoming of the published work on hyperacusis. Fabijanska *et al*.[7] undertook a postal questionnaire of tinnitus in Poland which included an unspecified question on hyperacusis. Of the 10 349 respondents, 15.2% reported hyperacusis (12.5% of males, 17.6% of females). Regional differences were also reported. A weakness of this report is the lack of specificity. More recently Andersson and co-workers[8] investigated the prevalence of hyperacusis in the adult Swedish population. Two methods were used—an internet study, wherein visitors to the website of a Swedish broadsheet newspaper were invited to complete a web-based questionnaire; and a postal population study. Of 1167 individuals who clicked upon the web banner 595 responded, a response rate of 52%. The point prevalence of hyperacusis in this group was 9%. The postal group comprised 987 individuals of whom 589 responded (response rate 60%) and the point prevalence was 8%. Participants were not asked if they had ever sought a medical opinion regarding their hyperacusis. Incidence data for hyperacusis do not seem to have been reported anywhere.

A coincidence of tinnitus complaint and of experiences of hyperacusis has been widely noted. Among patients attending tinnitus clinics with a primary complaint of tinnitus the prevalence of hyperacusis is about 40%;[9–11] and in patients with a primary complaint of hyperacusis the prevalence of tinnitus has been reported as 86%.[12] The apparent link has led to speculation about common mechanisms.[13]

Until recently it has not been possible to quantify the handicap associated with hyperacusis, but two instruments have now been published for this purpose. Khalfa *et al*.[14] describe data from a self-report hyperacusis questionnaire with 14 items 'normalized' on 201 individuals who had answered a recruitment advertisement. Principal component analysis indicated that three factors accounted for 48% of the variance—attentional, social and emotional. With a 27-item questionnaire examined in 226 patients with hyperacusis Nelting *et al*.[15] reached similar conclusions: 51% of the variance was accounted for by cognitive reactions, actional/somatic behaviour and emotional factors. This latter questionnaire is at present available only in German and neither has been shown to be sensitive to treatment effects, but such instruments do represent a step forward.

AETIOLOGIES

In the great majority of cases, no underlying medical condition can be found. The conditions in which hyperacusis has been reported as a symptom have been reviewed by Katzenell and Segal,[16] and those identified are listed in Box 1. It should be noted, however, that of the

Audiology (94), Addenbrooke's Hospital, Hills Road, Cambridge CB2 2QQ, UK
E-mail: dmb29@cam.ac.uk

peripheral conditions identified, several involve facial nerve dysfunction. Since the facial nerve innervates the stapedial reflex, which is a mechanism for reducing the perceived intensity of impulse sound, these conditions may reduce the efficacy of that reflex and hence increase the perceived intensity of sound. As such this does not meet a strict definition of hyperacusis.

What of the central conditions? Lyme disease is a systemic infection with the tick-borne spirochaeta *Borrelia burgdorferi* which targets specific body organs including the peripheral and central nervous systems.[17] Some caution must be exercised in interpreting reports of hyperacusis because facial palsy can be a feature, hence stapedial reflex dysfunction as described above. There are, however, reports of hyperacusis in Lyme disease without facial nerve dysfunction.[18]

Williams syndrome is a disorder characterized by deficits in conceptual reasoning, problem solving, motor control, arithmetic ability and spatial cognition,[19] with an incidence of 1 in 20 000 live births. As many as 90% of individuals with this syndrome report hyperacusis,[2] and a proposed mechanism is 5-hydroxytryptamine (5-HT) dysfunction[20]—see next section. Other conditions in which hyperacusis has been reported are middle cerebral aneurysm[21] and migrainous cerebral infarction.[22] A case series of hyperacusis in multiple sclerosis has been reported,[23] though the association is unusual.

Although most cases of hyperacusis are non-syndromic—i.e. do not reflect an underlying medical disorder—medical assessment is desirable.

MECHANISMS

Hyperacusis has several potential mechanisms which are not mutually exclusive; as with tinnitus,[24] the patient population is likely to be heterogeneous. The high prevalence of hyperacusis in Williams syndrome led Marriage and Barnes[20] to consider the mechanism in that condition and the extent to which it might be generalized to other individuals. Their suggestion that 5-HT might be implicated was based partly on the clinical observation that hyperacusis tends to occur in other conditions where 5-HT function is thought to be disturbed—namely, migraine, depression and post-traumatic stress disorder.[16,25] 5-HT does appear to have a role in modulating auditory gain and the determination of significance of sound.[26,27] However, there is no evidence that 5-HT disturbance contributes to hyperacusis of non-syndromic[28] types. Moreover, even in Williams syndrome the excessive auditory gain may be explained partly by the high incidence of otitis media with effusion and the associated conductive hearing loss.

Box 1 **Conditions associated with hyperacusis** (modified from Ref. 16)

Peripheral	Central
Bell's palsy	Migraine
Ramsay-Hunt syndrome	Depression
Stapedectomy	Post-traumatic stress disorder
Perilymph fistula	Head injury
	Lyme disease
	Williams syndrome

Sahley and Nodar[29] considered the observation that hyperacusis (and tinnitus) appear to increase in extent at times of tiredness, anxiety or stress. They hypothesize that, during stress, endogenous dynorphins are released into the synaptic region beneath inner hair cells. This might potentiate the neurotransmitter glutamate, causing sound to be perceived with excessive loudness. The model applies both to externally generated and to internally generated (tinnitus) sound, but empirical evidence in support has not yet been forthcoming.

Another potential mechanism is auditory efferent dysfunction. An auditory efferent system is common to all mammals, and in humans consists of both a lateral and a medial system. In the lateral system, whose function remains unclear, the pathways originate around the lateral superior olive and terminate on the primary afferent dendrite beneath the inner hair cell. In the medial system they begin medially with the superior olivary complex and terminate on the base of outer hair cells, and functions of the system appear to include modulation of auditory gain[30] and the behavioural response to sound (manifest in anatomical links with the reticular formation). Medial auditory dysfunction might contribute to both hyperacusis and tinnitus; thus, disturbance of the ability to modulate central gain might result in persistent sensitivity despite exposure to noise of moderate to high intensity. There is evidence against any such role, however, in that patients who have undergone vestibular nerve section (usually for symptoms of vertigo refractory to other treatments) do not complain of increased tinnitus or of loudness intolerance[31] and psychoacoustic testing of such patients reveals no decrement in auditory performance.[32]

For patients, hypersensitivity of hearing may evoke anxiety and even fear. This can be true for specific sounds or for sound in general. The links between the central auditory system and areas of the brain implicated in anxiety and fear are now under close scrutiny. Specifically, anatomical and functional links between the central auditory system and the amygdalae have been identified[33] (the amygdalae being an essential element of fear conditioning).[34] Such processes have been described as integral to the development of tinnitus-related distress, and also to the fear and anxiety component of hyperacusis. In view of the evidence that the central auditory system has a role in

setting auditory gain, the possibility of some central hyperexcitability should be considered. Jastreboff and Hazell[35] discussed this as a potential mechanism for hyperacusis. The experience of hyperacusis in patients with no apparent dysfunction or involvement of the peripheral auditory apparatus is circumstantial evidence in favour of this mechanism. Jastreboff and Hazell further speculate that such central hyperexcitability (manifest as hyperacusis) may represent a precursive state of troublesome tinnitus.

THERAPY

For many patients, the first reaction to hyperacusis is to protect themselves with ear plugs, muffs or other devices. There is, however, reason to believe that such strategies to decrease the intensity of sound entering the auditory system may further increase the central gain, exacerbating rather than improving the hyperacusis. In the past, patients had little choice but to resort to hearing protection devices since hyperacusis was not widely regarded as a genuine symptom. For tinnitus, tinnitus retraining therapy (TRT) was introduced in 1993,[35] and with minor modifications this has been advocated also for hyperacusis. After audiological and medical evaluation, the protocol[11] requires classification of the patient according to the tinnitus and hyperacusis state, and then 'directive counselling' about the auditory system, about mechanisms of tinnitus and hyperacusis and about the distress associated with them. Binaural sound therapy, from ear-level wide-band generators, is undertaken even when the symptoms are unilateral. Treatment is based on the notion of desensitization, and the sound intensity is increased from a low level gradually over time. No randomized controlled trials have been done on retraining therapy for hyperacusis; they would be hard to design in view of the twin elements of counselling and sound therapy. Several observational studies[36,37] have pointed to improvements in loudness tolerance, but the nature of training to do TRT (attendance at an examined course run by the originators) raises concerns about objectivity. Nevertheless, the approach taken by TRT practitioners—promoting understanding and insight and the use of low-level, non-threatening, wide-band noise—seems based on common sense.

For the psychological distress associated with tinnitus, cognitive–behavioural therapy (CBT) has been identified as the treatment of choice,[38] and this seems a reasonable strategy to counter the anxiety and stress associated with hyperacusis, together with information counselling, relaxation therapy and sound therapy. No evidence as to the efficacy of such an approach is yet available, and at present CBT therapists in the UK show little interest in tinnitus or hyperacusis. There is at present some tension between advocates of retraining therapy and advocates of psychological therapy, but the differences between the two are not great. Patients would probably benefit if the insights from both could be brought to bear.

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